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Baffle thrombosis in an adult with remote prior Scimitar vein repair mimicking massive pulmonary embolism

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DISCLOSURES

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KEY WORDS

Pulmonary embolus; artefact; computed tomography pulmonary angiography; Scimitar syndrome; Baffle thrombosis

ABSTRACT

A 58 year old man with a history of Scimitar Syndrome diagnosed and surgically repaired in early adulthood presented multiple times to the emergency department complaining of dyspnoea, chest pain and haemoptysis. Asymmetric pulmonary arterial flow rates between left and right lungs resulted in an apparent filling defect on computed tomographic pulmonary arteriography, which was repeatedly misdiagnosed clinically and radiologically as a massive pulmonary embolus. This case highlights the importance of understanding the pathophysiology and post-surgical complications of repaired congenital cardiovascular disease. Delayed phase acquisitions are often necessary to characterize the physiology of repaired congenital cardiovascular disease with associated shunts.

MAIN TEXT

Introduction

Scimitar syndrome describes a constellation of findings, most typically anomalous right pulmonary vein connecting to the right cardiac circulation (usually via the inferior vena cava), right sided pulmonary hypoplasia and systemic arterial collaterals supplying the right lung. Various additional cardiopulmonary variations and associations have been described including atrial (ASD) and ventricular (VSD) septal defects, pulmonary artery (PA) hypoplasia, diaphragmatic hernias, abnormal bronchopulmonary anatomy and sequestrations, vertebral and genitourinary abnormalities.[1,2]

The radiographic appearances of scimitar syndrome are well known and its accompanying asymmetric perfusion on conventional angiography is documented[3], however the associated appearance of abnormal pulmonary arterial blood flow on computed tomography pulmonary angiography (CTPA) has not previously been described. We report a case of post-operative scimitar repair and asymmetric pulmonary arterial perfusion with delayed right pulmonary blood flow, repeatedly misinterpreted as massive pulmonary embolus (PE).

Case Report

We describe the most recent clinical episodes of a 58 year old man, who presented to the Emergency Department (ED) of a metropolitan teaching hospital complaining of recurrent chest pain, haemoptysis and dyspnoea on four occasions with multiple intervening cardiac and respiratory outpatient clinics, before his diagnosis was confirmed on the fourth episode when he was transferred to a specialist cardiothoracic centre for evaluation. His course was complicated by a mis-interpretation of clinical findings and, in retrospect, misdiagnosis of PE during an unrelated presentation many years earlier.

1st encounter: the patient described a past history of single vessel ischaemic heart disease treated with right coronary arterial stent; paroxysmal atrial fibrillation; pulmonary hypertension; PE and

elective repair of Scimitar syndrome (right pulmonary veins redirected to left atrium via intra-atrial and trans septal baffle) discovered incidentally at age 29.

A triage chest radiograph (not shown) was performed on arrival. It demonstrated a previous median sternotomy, but no discernable scimitar vein remnant. Reduced right hemithoracic volume was misinterpreted as a pulmonary effusion and vague right sided parenchymal opacities were described. Initial history provided to the radiologist did not include the prior history of scimitar vein repair, but had been documented in the clinical notes, which were not requested by the reporting radiologist.

A CTPA study (Somatom AS+ 64 slice; Siemens Corporation, Munich, Germany) demonstrated a large proximal right PA filling defect (Figure 1a); right supraclavicular and mediastinal lymphadenopathy; right sided interlobular septal opacities, mosaic and nodular ground glass attenuation. A diagnosis of large right PA embolus was made with right pulmonary parenchymal changes attributed to the haemodynamic effects of large volume proximal obstruction.

As the patient was therapeutically anticoagulated for prior PE at presentation, concern for occult malignancy prompted whole body imaging (not shown), but no cancer was detected. Bilateral lower limb venous Doppler (not shown) was negative for deep venous thrombosis, but an inferior vena cava (IVC) filter was inserted and unfortunately complicated by a femoral insertion site arteriovenous fistula (treated conservatively). Six week follow-up CTPA (not shown) was unchanged.

2nd Encounter (5 months): A second ED presentation for recurrence of chest pain and haemoptysis prompted another CTPA, which demonstrated: improved, yet persistent asymmetric contrast opacification of the right pulmonary artery, stable appearing lymphadenopathy and parenchymal changes (Figure 1b). Despite this, and after apparent direct comparison with the initial CTPA, it was concluded that the PE had resolved.

3rd Encounter (11 months): Ongoing symptoms yielded another “positive” CTPA (not shown) with a diagnosis of refractory PE. Persisting right sided interstitial changes raised concern for lymphangitic carcinomatosis and a combined positron emission tomography (PET) CT study was performed (not shown) to exclude pulmonary artery sarcoma; it demonstrated intense FDG-uptake within the

lymphadenopathy, mild uptake in the right lung and no uptake in the right PA. A fine needle aspiration of the supraclavicular lymph node yielded only inflammatory cells.

4th Encounter (18 months): Suspicion of an alternative diagnosis after another apparently positive CTPA (not shown), prompted a contemporaneous CMR (Magnatom Aera; Siemens Corporation; Munich, Germany). Axial T1 post-contrast sequence demonstrated no PA filling defect (Figure 2a). The multi-phase dynamic magnetic resonance pulmonary angiography (MRPA) resolved the ongoing diagnostic dilemma.

Early phase MRPA (Figure 2b) confirmed asymmetric PA perfusion hemodynamics resulting in delayed contrast flow artifact in the right main PA, mimicking a large proximal PE. Late phase MRPA sequences (70 seconds) demonstrate that delayed acquisition permitted contrast to opacify the more slowly filling right PA system (Figure 2c), accounting for the apparent resolution of PE (Fig 1b), which in retrospect was due to a late CTPA acquisition (note the opacification of the descending thoracic aorta). No flow through the intra-atrial baffle could be demonstrated.

Outcome

The patient was diagnosed with baffle occlusion and right pulmonary venous hypertension which was a significant contributor to the reduced right pulmonary arterial flow. The IVC filter was removed without incident and anticoagulation was ceased. At the time of writing the patient reports no subsequent episodes of hemoptysis and as such has not yet required embolization of his arterial collaterals. His chest pain and dyspnoea persist but have improved.

Discussion

Clinical manifestations of Scimitar syndrome are variably dependant on the extent of left-to-right shunt, presence of ASD or VSD, valvular abnormalities and degree of pulmonary hypertension.[4] Perfusion of the right lung is commonly reduced in Scimitar syndrome,[5] in both pre-operative and post-operative states.[6] Perfusion abnormality is likely the result of hypoplastic lung, a small right PA or pulmonary venous obstruction, and whilst surgery can correct the vascular shunt, underlying pulmonary architectural abnormalities persist. These background abnormalities can result in

lymphatic congestion, which would explain the persistent parenchymal findings in the right lung in this case (Fig 1c) rather than the erroneous suspicion of lymphangitic carcinomatosis.

A literature search (Pubmed/MEDLINE, CINAHL and Google Scholar) yielded only one publication with images demonstrating such asymmetric perfusion on (conventional) pulmonary angiography in the setting of Scimitar Syndrome.[3]

In almost every case, the CT studies were read by different radiologists and the history of previous PE may have prejudicially resulted in an element of *confirmation bias*. Interestingly, the previous diagnosis of PE was made many years earlier by interstate intensive care physicians during an admission for a severe community acquired pneumonia. The conclusion was drawn, despite a negative CTPA (presumably performed on an older slower machine) and was based on asymmetric pulmonary arterial pressures during invasive monitoring. Unfortunately this history, potential bias and misinterpretation resulted in a total of six CTPA's, one CT chest/abdomen/pelvis and one PET/CT performed during the described timeframe, with an estimated effective dose of 83.4mSv[* 7,8].

CTPA has replaced conventional pulmonary angiography to become the gold standard in the diagnosis of PE, but has inherent limitations. Whilst it is essentially non-invasive and can provide excellent anatomic detail, it does not, under typical circumstances provide dynamic information about blood flow through the pulmonary vascular bed.

Current multi-detector CT technology enables high speed acquisitions reducing respiration and motion artifact, significantly improving the image quality and reducing the number of indeterminate studies. However, one shortcoming is the potential to capture unusual flow related phenomena

[*] Assuming average estimated effective doses of 7.9.mSv (CTPA), 21.5mSv (CT chest, abdomen & pelvis)[7] and 14.5mSv (PET/CT)[7].

related to abnormal (delayed) pulmonary hemodynamics. Such appearances are obvious in a dynamic setting such as conventional pulmonary angiography, however single phase acquisition of CT such occurrences have the potential to be misleading. Older CT technology had longer acquisition times, enabling more even contrast mixing and distribution, even throughout abnormal vascular beds. Such flow anomalies have been reported to mimic pulmonary emboli in the setting of single cardiac ventricle,[9] Fontan procedure[10] and iatrogenic pulmonary vein stenosis post-cardiac radiofrequency ablation.[11]

The complicated imaging appearance and concerning clinical findings of this episode took almost 18 months to be resolved, but concluded a course of missed diagnosis and incorrect management lasting many years. Asymmetric PA opacification has various unusual and potentially confusing causes (Table 1) [9, 10, 11, 12, 13, 14, 15, 16]. The asymmetric PA flow artifact was repeatedly misinterpreted by many experienced radiologists and clinicians. Apart from poor opacification of the right pulmonary artery, inhomogeneity of the contrast pool within the left and main pulmonary arteries was an additional clue to the diagnosis which was not identified. Only one CTPA report included a description of post-operative scimitar syndrome, however, the link was not made to the perfusion abnormality. Whilst MRI was available, limited access to local specialist cardiothoracic radiology opinion may have resulted in delay in request for CMR.

Failure to recognize the background pathophysiology and complications of corrected congenital cardiovascular disease led to the mistaken diagnosis and extensive treatment (with complications) of PE as well as suspicion of malignancy.

CMR is the gold standard in dynamic cardiovascular investigation, particularly for flow related abnormalities. Whilst CMR tends not to provide the spatial resolution of CTPA, it can provide dynamic acquisitions enabling assessment of perfusion, flow direction and flow rates without exposing the patient to radiation or femoral puncture. It should be sought early in the diagnostic pathway for complex questions particularly post-operative congenital cardiopulmonary cases.

Table 1 - Causes of asymmetric opacification of the pulmonary artery and PE mimics (from Remy-Jardin et. al.[13] supplemented with [9 - 12, 14- 16])

Unilateral pulmonary artery obstruction of extrinsic, mural, or endoluminal origin
Unilateral increase in pulmonary vascular resistance, secondary to: Proximal or distal bronchial obstruction Bronchiolar obstruction Parenchymal destruction Extensive air-space consolidation Pleural restrictive or expansive process Congenital anomalies (e.g. bronchopulmonary sequestration, pulmonary artery absence/hypoplasia) Conditions with elevated venous pressure (e.g. iatrogenic pulmonary vein stenosis)
Hemithoracic shunting of blood from systemic to pulmonary circulation following surgical anastomoses of Blalock, Waterston, Potts, Glenn and Fontan (single cardiac ventricle)
Inspiration associated artifact (transient interruption of the pulmonary arterial contrast column)[a]
Respiration movement artifact[a]
Pulmonary arterial catheter[a]
Partial volume artifact[a]

[a] - Specific to computed tomography

Summary

This case highlights the importance of understanding the pathophysiology and post-surgical complications of repaired congenital cardiovascular disease, as well as an awareness of the challenges faced when interpreting diagnostic imaging studies with potentially confusing artifacts. When identified, significant asymmetric pulmonary arterial flow always requires an explanation – the use of delayed phase acquisitions is often necessary to characterize the physiology of repaired congenital cardiovascular disease with associated shunts. Early involvement of subspecialty radiologists should be sought in complex cardiothoracic cases.[17, 18]

FIGURE CAPTIONS

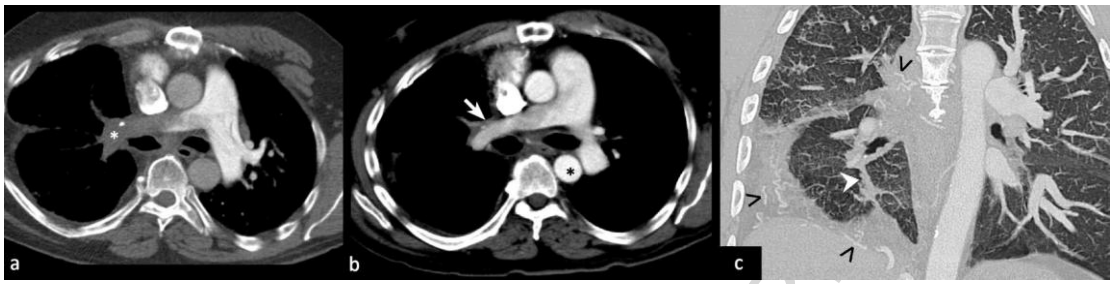

Figure 1a - Large right PA filling defect (white asterisk), with relatively dense contrast opacification of the left pulmonary arterial tree.

Figure 1b – Improved contrast opacification of the right PA (white arrow), however disparity between left and right pulmonary flow remains. Dense contrast opacification of the descending aorta (black asterisk) reflects unintentional late acquisition.

Figure 1c - Network of arterial (bronchial and hepatic) collaterals supplying the right lung (open black arrowheads). Post-surgical remnant of the Scimitar Vein (white arrow head).

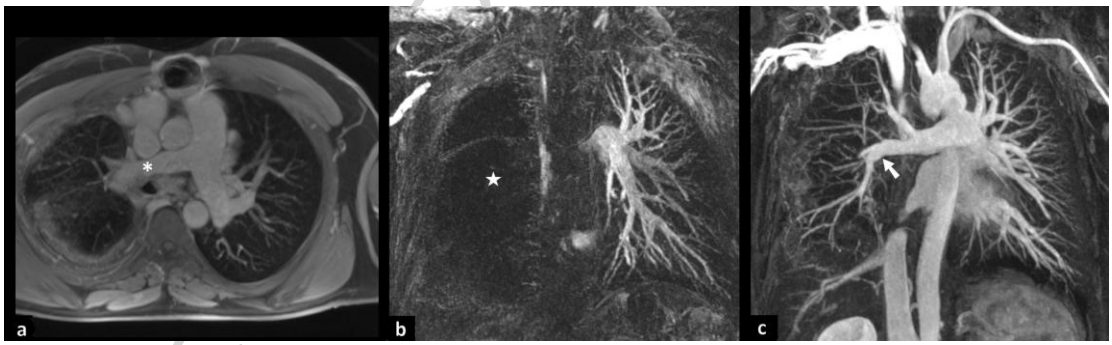

Figure 2a – The apparent right PA filling defect is not present on post contrast (90 sec) T1 fat-suppressed VIBE (3D Volumetric interpolated breath-hold examination) sequence MRI of the thorax (white asterisk).

Figure 2b - Coronal MIP MRPA - early phase (30 sec) acquisition demonstrates absent right PA flow (white star) corresponds to CTPA findings in Fig 1a.

Figure 2c - Coronal MIP MRPA late phase (70 sec) acquisition demonstrates delayed right PA flow (white arrow) corresponds to CTPA findings in Fig 1b.

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